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Retiform Hemangioendothelioma

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Summary

A 30-year-old female presented with multiple, erythematous, velvety, slowly growing, asymptomatic nodules on the scalp of two-year duration. Biopsy revealed a vascular tumor with arborizing blood vessels arranged in a retiform pattern, lined with hobnail endothelial cells that stained positively with CD34, with few intravascular papillae. Two year follow up of the case after surgical excision showed multiple recurrences but with no metastasis.

Introduction

Retiform hemangioendothelioma (RH) is a rare, recently described variant of low-grade angiosarcoma, characterized by indolent clinical behavior and closely related to Dabska's tumor (DT). Calonje et al. first described the tumor in 1994[1].

Most of the tumors presented within the second to fourth decades of life, with no sex predilection. Most reported cases were seen in the upper and lower limbs [1, 2, 3, 4].

RH usually occurs as a single lesion, but multiple lesions affecting different anatomical sites were reported afterwards[2].

Case Presentation

A 30-year-old female presented with a 2-year history of multiple rose-colored nodules of velvety surface, on the scalp and nape of the neck. The nodules were slowly increasing in size and number. Lesions were asymptomatic apart from discomfort upon combing the hair, and occasional bleeding. There were no other complaints or physical signs, nor was there any local lymph node enlargement (Fig 1).

A 6 mm punch biopsy was taken, and histopathological examination revealed numerous dilated vascular spaces in the lower dermis, showing very characteristic arborizing blood vessels, arranged in a retiform pattern [reminiscent of normal rete testis (Fig 2)].

These blood vessels were lined by monomorphic endothelial cells, with prominent apical nuclei, and scanty cytoplasm. These cells are described as having a hobnail or matchstick appearance. Occasional intravascular papillae with hyaline collagenous cores can be seen in some of these vessels (Fig 2). Staining with CD34, which is an endothelial marker, was positive, restricted to the intravascular endothelial cells.

Our final diagnosis was thus retiform hemangioendothelioma.

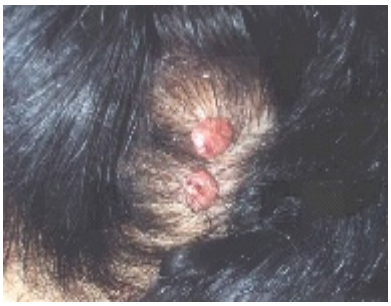

Figure 1

Retiform hemangioendothelioma: multiple lesions on the scalp.

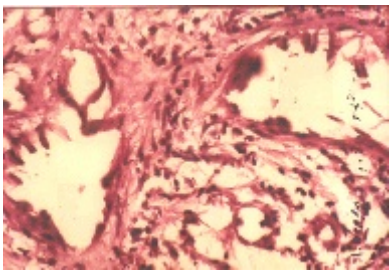

Figure 2

H&E section: numerous dilated vascular spaces in the lower dermis, showing very characteristic arborizing blood vessels, arranged in a retiform pattern

Discussion

The term low-grade angiosarcoma refers to a group of vascular neoplasms that have a histopathological appearance intermediate between haemangioma and angiosarcoma. This group includes epithelioid hemangioendothelioma, endovascular papillary angioendothelioma (Dabska's tumor), and retiform

hemangioendothelioma[5].

In the 15 cases of RH, described by Calonje[1], 6 tumors arose on the lower limbs, 4 on the upper limbs, 3 on the trunk, and 1 each on the penis and scalp. Our case is then the second case to be reported to occur on the scalp.

RH usually occurs as a single lesion. Reports of multiple lesions affecting different anatomical sites were reported afterwards[2]. Our case is the first case to show multiple lesions affecting one anatomical site.

The tumor has non-specific clinical features, it may occur in the form of a slowly growing exophytic mass, a plaque-like lesion, or a dermal or subcutaneous nodule. All these presentations are misleading and do not suggest the vascular nature of the tumor[6]. However microscopically it shows very characteristic findings in the form of arborizing blood vessels arranged in a retiform pattern, and lined by monomorphic hobnail endothelial cells, together with occasional intravascular papillae with hyaline cores[1, 2, 3, 4].

Immunohistochemically, the tumor cells react with endothelial markers; CD31, CD34, Factor VIII related antigen, and bound ulex europaeus agglutinin[11, 3]. In our case, reaction of the tumor cells to CD34 was seen restricted to the intravascular endothelial cells.

Multiple recurrences are common, but metastasis has so far been reported in only one case, and that is why it is considered a low-grade neoplasm, and there have been no tumor-related deaths[7].

RH and DT share some common biologic behavior and histologic features. Many authors believe or propose that RH is the adult variant of DT. DT, which mostly occurs in children, shows no retiform architecture, is composed of interconnecting cavernous vascular spaces resembling lymphatics, contains more intravascular papillary projections with central hyalinized collagenous cores and the hobnail endothelial cells are mainly seen in the vessels near the surface[8, 9].

The most important differential diagnosis of RH is angiosarcoma, which is of great importance for therapeutic and prognostic reasons; however, angiosarcoma presents in a different clinical setting, shows cytologic atypia and mitosis, shows dissection between collagen bundles, and absence of hobnail endothelial cells [10, 11].

Being a low-grade malignancy, RH needs less aggressive treatment lines. Wide surgical excision is enough, but should be followed up for recurrences[6]. In our case, surgical excision of the tumors was done, and follow-up of the case continued for two years, with many recurrences, but no metastasis was seen.

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